A rare cause of intestinal hemorrhage: Stromal tumor of duodenum

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ABSTRACT
Stromal tumor of the duodenum is rarely seen among the gastrointestinal stromal tumors. In this paper, a patient presented with obscure gastrointestinal bleeding and the small bowel endoscopy revealed a bleeding ulcer at the second part of duodenum. The patient underwent segmental duodenectomy. The histopathology of the mass confirmed the diagnosis of duodenal stromal tumor. [Turk J Cancer 2004;34(4):163-165]

INTRODUCTION
Gastrointestinal stromal tumor (GIST) is the most common mesenchymal tumor of the gastrointestinal tract (GI) (1). In the past years, most of these tumors had been referred to as leiomyomas, leiomyoblastomas and leiomyosarcomas (2). Stromal tumor term was first described by Schaldenbrand and Appelman in 1984 (1). GISTs arise most commonly in the stomach (60-80%), followed by the small intestine (40%) and then the colon, rectum or esophagus (3). The age range is generally 40 to 80 at the diagnosis. GISTs arise rarely in the duodenum and clinical presentation is variable. A palpable abdominal mass is the most frequent sign. A half of GIST is silent until it reaches a large size, at which point it may cause acute massive hemorrhage into the intestinal tract or peritoneal cavity from tumor rupture so second most frequent sign is GI hemorrhage. Obstructive jaundice may be seen if GIST is located near the ampulla Wateri (4). Small asymptomatic lesions arising in the duodenum is found incidentally during laparatomy for other reasons (5). Surgery remains the standard treatment for nonmetastatic GISTs (6,7). The tumor may have a pseudocapsule and should be removed with total duodenectomy or partial duodenum resection. Complete resection of GIST isn’t curative as recurrence is quite common. In patients with local disease, the recurrence rate is 35% (1). A major diagnostic criterion of GISTs is expression of kit (CD117) and additional criteria include CD 34 and desmin (8,9).
In this report, we reviewed the diagnosis, pathology and treatment of a patient presenting with acute hemorrhage from duodenal GIST.

**CASE REPORT**

A 61 year old man had hematemesis and melena of 2 months duration. He had four bleeding episode in last 2 year. The endoscopic examination demonstrated a bleeding ulcer in the junction of second and third part of duodenum. Sclerotherapy was ineffective in both attempts. Computed tomography (CT) demonstrated that there was a rounded tumoral mass near the descent part of duodenum and multiple nodular lesions of the liver. The tumor was located on the second and third part of duodenum. The tumor size was 5 cm. Under this circumstances the patient underwent laparotomy. On exploration we encountered a tumoral mass which was originated in the second and third part of the duodenum wall. Tumor didn’t diffusely infiltrate the retroperitoneum, it tended to hang from adjacent structure. The patient underwent segmental duodenectomy with end to end anastomosis. The macroscopic appearance of the 5 cm tumor showed submucosal localization and mucosal ulceration. In cut section the tumor was grey-whitish colored, rubbery with necrotic and cystic degeneration in same areas (Figure 1). The histopathology of the tumor composed of interlacking bundles of spindle cells without mitosis and pleomorphism. Immunohistochemical staining with CD117 was found diffuse positive but other markers for CD34, desmin, Ki 67 were negative (Figure 2).

**DISCUSSION**

GISTs are a heterogenous group of diseases that differ in frequency, clinical symptoms and pathologic behavior in patients. Until recently it was categorized as a tumor of smooth muscle derivation. In the mid 1980s GIST has been suggested as a distinct entity.

When localized GIST lesions tend to be primarily intramural and submucosal, often evading clinical diagnosis unless they rupture and bleed. Duodenal stromal tumor presents different clinical symptoms. One of the most important of them is acute hemorrhage into the intestinal tract due to necrosis of mucosa. It may be seen in up to 25% of GIST patient. GI bleeding can stem from ulcerating benign or malignant tumors.

Several radiological techniques are used to image primer GISTs and metastatic lesions. These include double-contrast GI X-ray series with barium, endoscopic ultrasonography, computed tomography and magnetic resonance imaging (MRI) (4). Recently positron emission tomography scanning has been shown to be more sensitive than MRI.

When GISTs are initially diagnosed, approximately 25% are frankly malignant. The criteria for this designation include metastasis or an invasion to adjacent organs. It is often difficult to predict the clinical behavior of GIST. Many pathologists believe that none of GISTs are truly benign. So terminology of the GISTs are being replaced “low risk” versus “high risk”. Prognostic factors for high risk are large tumor size (> 5 cm), many mitotic figures (> 5 per 50 HPF), fast growing and often infiltrating as in our case.

The accurate diagnosis of GIST requires IHC staining for c-kit (CD 117) expression. Other antigenic markers...
(CD34, SMA, Desmin 2, S 100, Ki 67) are also variably positive.

Surgical resection has been the mainstay of therapy for GIST. Negative pathologic margins of resection generally is not difficult because GIST tends to hang from, not diffuse infiltrate, the organ of origin. Tumor rupture before or during resection is predictor of poor outcome. Local peritoneal tumor seeding is common. In a study, many patients (40% in twenty four months) develop recurrent GIST despite complete resection of their primary tumor (10). The liver is the most common site of recurrence. Treatment options for recurrent and metastatic diseases have been limited. In our case, we did not perform liver resection due to poor general condition. Clinical trials have shown that patients benefit from Imatinib therapy, so Imatinib was used in our case (11).

In conclusion a high level of suspicion to detect this malignant tumor especially in patients presenting with massive gastrointestinal bleeding and effective surgical treatment allow better clinical outcome.

References